

SICKLE CELL ANEMIA IN CHILDREN: A MORBIDITY PROFILE IN A TERTIARY CARE HOSPITAL



Paediatrics

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ABSTRACT

To study the commonest clinical complication requiring admission of children with sickle cell disease. Indication of Blood Transfusion received by the patient. To assess the vaccination status which also includes special vaccines like Pneumococcal, Typhoid, Meningococcal along with universal immunisation programme vaccines. It was a retrospective case series study which was conducted at tertiary care hospital NKPSIMS and LMH, Nagpur from July 2013 to June 2014. The case records were analysed as per the pro forma. Total of 68 case records were studied of which 11 case records were excluded due to incomplete data. Total of 57 subjects were included in the study and data was analysed. Result- Fever (35%) constituted as a sole presentation for admission. The commonest indication for Blood Transfusion were Stroke (7%) and Severe Malaria (7%). No statistical significance was observed regarding blood transfusion in febrile patients. Only 10.5% of subjects were vaccinated with Pneumococcal vaccine and 28% of subjects were vaccinated by Typhoid vaccine. Conclusion: Even with the widespread availability of special vaccines for children with Sickle Cell Anemia, there is a need for awareness for the beneficiaries which can be achieved by utilising peripheral workers like Anganwadi & ASHA workers and also use of media in regional languages.

KEYWORDS:

Sickle-morbidity-mortality-hydroxyurea- vaccination

INTRODUCTION

Sickle cell disease (SCD), causes a wide range of severe and even life threatening consequences, caused by a single base pair change thymine for adenine, at the 6th position of Beta globin molecule, a protein vital for carrying oxygen in the blood. As a result of this mutation, individuals with SCD experience lifelong complications including anemia, infections, stroke, tissue damage, organ failure, intense painful episodes, and premature death. Prevalence of sickle cell trait varies greatly between different regions wherein it could be upto 40 percent in some areas of sub-Saharan Africa, eastern Saudi Arabia, and central India[8]. In developing countries maximum children with SCD do not survive to adulthood.

Africa has three fourth of the global sickle burden, more than half of these affected children die before the age of 5, primarily due to infections – pneumococcal, other bacterial, and malaria[10].

In India, sickle cell disease (SCD) is common in Vidarbha, Chhattisgarh, Madhya Pradesh, Orissa, Gujarat, Tamil Nadu and Andhra Pradesh[2].

Presence of splenomegaly and preserved splenic function are important for transmission of pneumococcal infection. Risk of Invasive pneumococcal disease falls sharply after the age of five years and a functioning spleen adds to the benefits of decreased infection[9]. Keeping these factors in mind introduction of hydroxyurea early after diagnosis and effective vaccination in all sickle cell patients can significantly increase survival of our future generation.

Aim and Objectives of the Study:-

The study was conducted with following objectives in mind:

1. To study the commonest clinical complication requiring admission of children with sickle cell disease
2. Indication of Blood Transfusion received by the patient
3. To assess the vaccination status which also includes special vaccines

like Pneumococcal, Typhoid, Meningococcal along with universal immunisation programme vaccines.

Material & Methods:

1. Place of Study: This study was conducted at central India Nagpur (Maharashtra)
2. Period of Study: July 2013 – June 2014 (11 month)
3. Inclusion Criteria:
 1. All paediatric patients up to the age of 18 years of age diagnosed with sickle cell anaemia admitted from July 2013 to June 2014 were enrolled in the study.

2. The case records were analyzed in detail for the cause of admission.

4. Exclusion Criteria:

1. Patients discharged within 24 hrs of admission to hospital were not included.

4. Sample size: All paediatric patients up to the age of 18 years of age diagnosed with sickle cell anaemia, referred at Medical College NKPSIMS & LMH, Nagpur were included in the study (N=57)

5. Study Design: Retrospective case series study

6. Methodology:.

- The case records with relevant data were studied as per the proforma and later analyzed by using appropriate statistical tests.

7. Statistical Analysis

Incomplete or inadequate supporting data to the diagnosis mentioned were excluded from the study. The case records with relevant data were studied as per the proforma and later analyzed by using percentage analysis, Mean, Standard Deviation, t-value of significance of mean differences in term of various variable. We have entered all data in Microsoft Excel and further Statistical Analysis was done with the help of QI-Macros 2014 Software.

Results

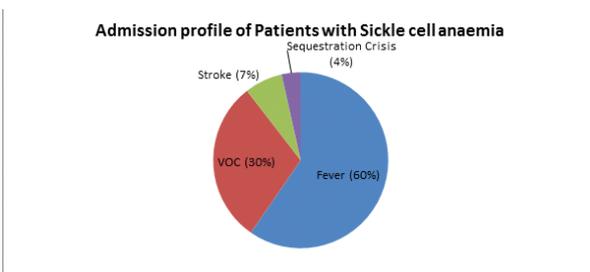
Amongst the causes of admission, fever contributed to the maximum cases (60%). Other causes were VOC (30%), Stroke (7%) and Sequestration Crisis (4%) for the cases.

Table no. 1:1 – Admission profile of patients with sickle cell anaemia

DIAGANOSIS	Total no of Clients	Percentage (%)
1. Fever	34	60 %
2. Vaso-occlusive crisis (VOC)	17	30 %
3. Stroke	4	7 %
4. Sequestration Crisis	2	4 %

% Percentage

Figure no. 1:1 – Admission profile of patients with sickle cell anaemia



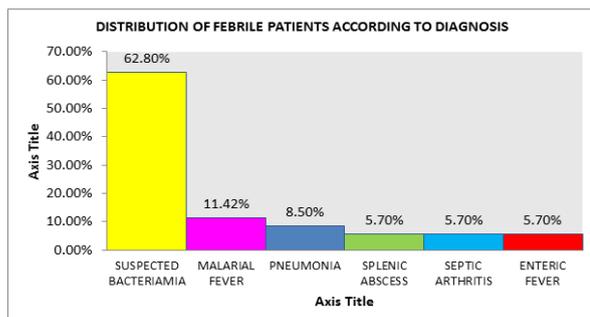
All the patients of Sickle Cell Anemia admitted with fever were scrutinized for the etiology of which 62.8% were because of suspected bacteraemia whereas 11.42% were of malarial fever, 8.5% of Pneumonia & 5.7% each of Enteric Fever, Splenic Abscess and Septic Arthritis respectively.

Table no. 1:2 – Distribution of febrile patients according to diagnosis

Sino.	Diagnosis	Percentage (%)
1	Suspected Bacteraemia	62.80 %
2	Malarial Fever	11.42 %
3	Pneumonia	8.50 %
4	Splenic Abscess	5.70 %
5	Septic Arthritis	5.70 %
6	Enteric Fever	5.70 %

% Percentage

Figure no. 1:2– Distribution of febrile patients according to diagnosis



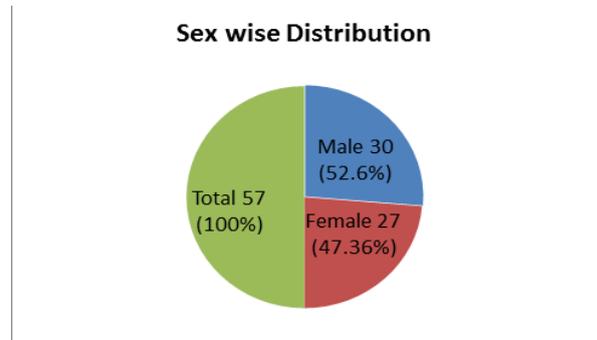
% Percentage

Table no. 1:3- Sex wise distribution

Sex wise distribution		
	No of patients	Percentage
Male	30	52.6%
Female	27	47.36 %
Total (n)	57	100

% Percentage

Figure no. 1:3- Sex wise distribution



% Percentage

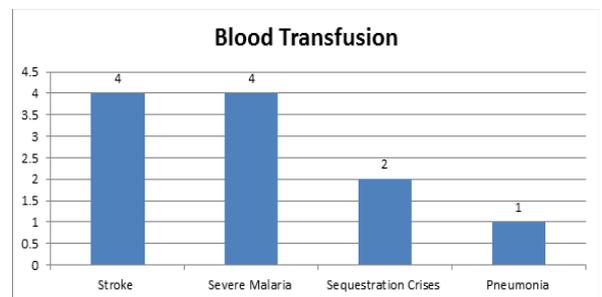
Out of 57 sickle cell patients only 11 needed blood transfusion i.e 19.29%. (See table no. 1:4). Commonest indication was Stroke and Severe Malaria which were 7.01% respectively. Other indications were Sequestration Crisis 3.5% & 1.75% of children with Pneumonia received blood transfusion. (See table no.1:4)

Table no. 1:4- Patients needed blood transfusion

Sino	Indications for Blood Transfusion were:	No. of Patients	Percentage
1	Stroke	4	7.01 %
2	Severe malaria	4	7.01 %
3	Sequestration Crises	2	3.50 %
4	Pneumonia	1	1.75 %

% Percentage

Figure no. 1:4- Patients needed blood transfusion



Non- significance on comparison of blood transfusion in febrile and afebrile patients was observed. The statistical analysis was done using t- test.

Table no. 1:5- Blood transfusion received in patients of sickle cell anaemia

Sino	Blood transfusion	Total Score Mean (m)	Standard Division (SD)	t- value	Significant Level
1	Febrile Patients	3.437	1.931	-1.357	Non-significant at P>0.05
	Afebrile Patients	3.968	3.386		

Analysis for vaccination status in children about the following vaccines namely Hib, Typhoid, Pneumococcal and Meningococcal was done and it was observed that 73.6% subjects received Hib vaccine whereas 28% received Typhoid and 10.5% received Pneumococcal vaccine. None of the subjects received Meningococcal vaccine. (See table no.1:6)

Table no. 1:6- Vaccination status

Vaccination Status		
Sino.	Vaccination	Percentage (%)
1	HIB	73.6%
2	TYPHOID	25%
3	PNEUMOCOCCAL	10.52%
4	MENINGOCOCCAL	0
5	INFLUENZA	0

% percentage

Table No. 1:7- Showing patients on hydroxyurea

On Hydroxyurea	Not on Hydroxyurea	Total
18 31.5 %	39 68.5%	57 100%

Conclusion

Fever is the most common etiology followed by Vaso-occlusive crisis (VOC) requiring admission to the hospital. Amongst the causes of fever, Suspected Bacteremia is the most common reason which should be thought and treated aggressively to prevent disease related complication.

Present study showed that blood transfusion is commonly given in children with stroke and sequestration crisis, both belonging to non-infective etiology. In endemic areas malarial fever with complications requires blood transfusion as per the treating clinician's discretion. Stroke is also a significant complication in SCD, with the potential for major morbidity and mortality [12]. In HbSS disease, the incidence of overt stroke is 11% by age < 20 years. [13] In present study statistically no significance was noted for blood transfusion in febrile and afebrile patients. Overall awareness for vaccination and hydroxyurea should be emphasized for better outcomes in children suffering from Sickle cell anaemia. The present study shows low percentage of vaccination and use of hydroxyurea amongst patients of sickle cell anaemia.

Thus, vaccination in paediatric sickle cell disease patients represents an important aspect of their preventive care [14]. At age 2 and 5 years, paediatric patients should receive Pneumococcal vaccine and also receive quadrivalent meningococcal vaccine at age 2 years as the Advisory Committee on Immunization Practices (ACIP) recently recommended vaccination starting at age 2 years for populations at increased risk of invasive meningococcal disease. [15]. Annual influenza vaccination is recommended because paediatric sickle cell disease patients are more likely to require hospitalization for influenza-related complications and experience more complications from influenza infections than children without sickle cell disease [16]. A double-blind randomized controlled trial of hydroxyurea in adults revealed lower rates of painful crises, ACS and unscheduled blood transfusions in patients treated with hydroxyurea. These results were recently confirmed in a multi-centre, double-blind, randomized controlled trial of hydroxyurea in young children with SCD (BABY HUG).

Delimitation of the research

- Limited sample size
- Areas based research

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